A FURTHER REPORT

ON THE

REMARKABLE SERIES OF CASES OF MOLLUSCUM FIBROSUM IN CHILDREN

COMMUNICATED TO THE SOCIETY BY DR. JOHN MURRAY IN 1873[1]

BY

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Our reasons for further referring to these cases are, first, that, owing to the death of one of them in Islington Infirmary, we were able to obtain a supply of material for examination from different parts; secondly, that we had also the opportunity of taking illustrative photographs to show the further development; and thirdly,

[1] 'Transactions of the Royal Medical and Chirurgical Society,' vol. lvi, p. 235.

that, owing to the fact that these cases appear to be unique, it may be of interest to the Fellows of this Society to have their attention again drawn to them.

As thirty years have passed since the able and exhaustive report of Dr. Murray upon these cases, it may be well, perhaps, to very briefly sketch their history. From Dr. Murray's report, and from the verbal communication of a surviving brother, we have found that the father and mother of the patients were cousins on the paternal side, and that the eldest and youngest children of the family, who did not suffer from the disease, were born under

FIG. 1.

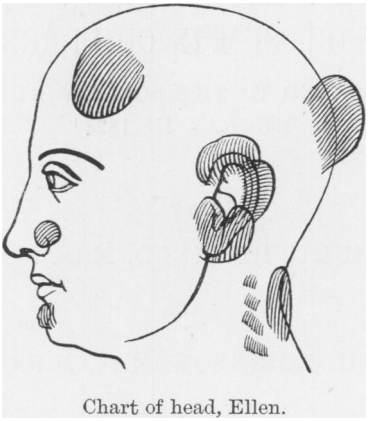


Chart of head, Ellen.

better hygienic conditions than three intermediate children, all of whom suffered from this peculiar condition.

Of these three children we have been able to see two, namely, Ellen, the eldest, who resides at present in the Putney Home for Incurables, and Richard, who died in the Infirmary, and from whom our photographs and material were obtained. We wish to acknowledge our indebtedness to the courtesy of the Matron and Secretary of the Putney Home in allowing one of us to examine Ellen. The third child has, we believe, been dead some time, and we were unable to find more about her than is

contained in Dr. Murray's report. From this report it appears that in the case of Ellen the disease was first noticed at the eruption of the teeth, and did not appear on the skin until she was two years old, while in Richard the gums were observed to be diseased at three months, and the skin at six months.

In reporting the further progress of the two patients whom it was our privilege to see, it will be better to take the female first, as it is to her case that Dr. Murray devotes the greatest attention in his report. From this

FIG. 2

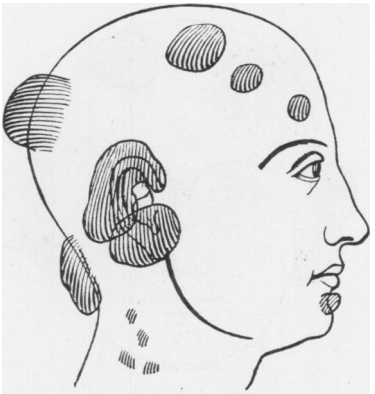


Chart of head, Ellen.

report, and the admirable accompanying illustration, we gather that there has been an increase in the size of the tumours rather than in their actual number, though we find several noted on our charts in positions which are not mentioned by Dr. Murray. Most of the tumours in the previous report were from a pea to a walnut in size; in fact, this latter is the greatest size mentioned. Our charts of the present condition show that the growth of tumours has progressed to such an extent that the patient, instead of presenting a somewhat curious aspect, is now so hideously deformed that she wears her head constantly

bandaged up. The tumours have now the following appearance:—On the forehead there are four, three on the right and one on the left side, that on the left side being about the size of an orange, and all being, as are almost all her tumours throughout, of a deep plum-colour. The ears are so deformed that it is impossible to make out accurately the original anatomical landmarks, but on both sides the aggregated lobed tumours reach a mass the size of the fist. On the left ala nasi and on the chin single tumours are present. The germ of that on the nose is figured in Dr. Murray's illustration, while that

FIG. 3.

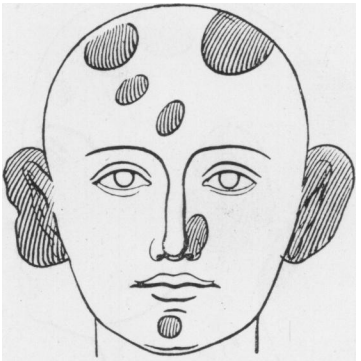


Chart of face, Ellen.

shown in his picture on the right ala nasi has apparently disappeared. He calls it a verruca plana, and this might suggest that it was not of the same permanent character as the other tumours; but he also describes that on the right side of the nose as similar, and this is now undoubtedly one of the fibromatous class. The gums apparently remain in much the same state as when he saw them, being mostly covered with small pea-sized fibromata. There is one large tumour on the occipital region, one smaller one, and a double row of quite insignificant ones running down the back of the neck. A large growth, the size of half an orange, lies between the scapulæ, and a few smaller

ones lie about it. The front of the trunk is practically clear. On the extensor surfaces of both elbows there are large diffluent and lobulated tumours, somewhat similar

FIG. 4.

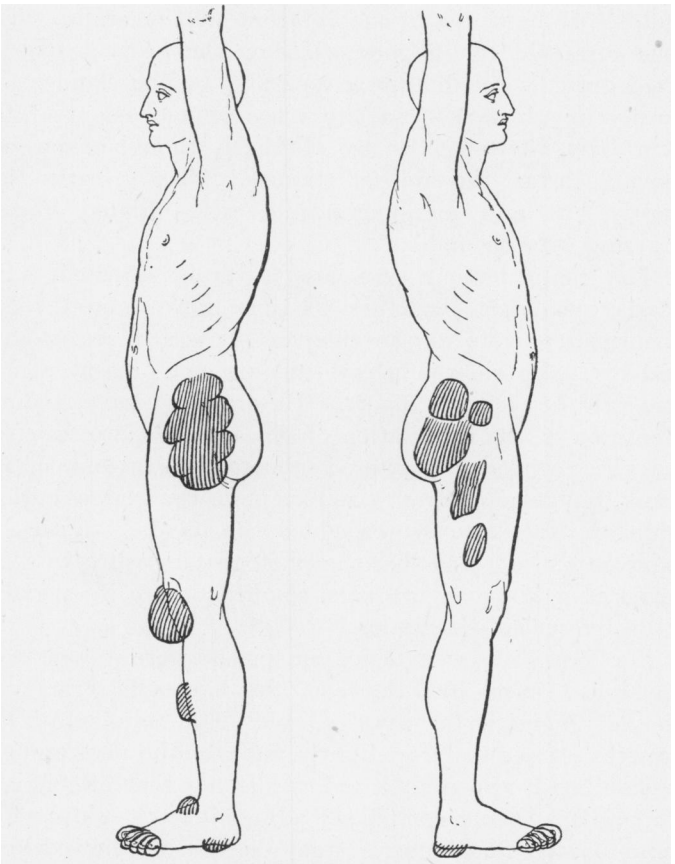


Chart showing distribution of tumours of lower extremities in Ellen.

growths over the extremities of both ulnæ, and the whole of the ends of the fingers of both hands are wrapped in lobed bluish tumours, much resembling those shown in the original picture. The gluteal region is almost entirely

occupied by huge, soft, lobed tumours extending from nearly as high as the posterior superior iliac spine into the upper thirds of the thighs. The trochanter regions are similarly occupied, as is the front of the left knee. There are a couple of growths on the left leg, one in the middle of the shin and one in front of the ankle. The toes resemble the fingers, while on the heels are good-sized growths not finding any parallel on the hands. In hardly any instance can any trace of movement of the skin over the growths be obtained, though they are movable over the deeper tissues. The growths are mostly soft and semi-fluctuating, with dilated vessels coursing over them.

The male patient was less severely affected. His hands, which in Dr. Murray's time only showed slight abnormality, were at the time of his death remarkable, and his scalp showed many tumours not present at the time of the former report. In order to save tedious repetition of the localisation of the tumours, we may say that there were some scattered subcutaneous tumours on the trunk, but none of great size except on the gluteal region, while the feet exactly resembled the hands. With this explanation we think that the photographs will give a far better idea of his unfortunate condition than we are able to do by verbal description.

For purposes of examination pieces were taken from the lobed nasal growth, from the ear, and from one of the fingers. It may be said that, clinically, the growths seemed to differ slightly, those on the nose and ear resembling in appearance and consistency that of ordinary rhinophyma, being completely adherent to the skin, while those on the fingers were stony hard and allowed very slight movement of the skin over them.

With the increased knowledge of the disease formerly known as molluscum fibrosum, and now called neuro-fibromatosis, it became necessary to examine for the presence of nerve-fibres. Specimens were, therefore, specially hardened for the Weigert process, and, owing to the

kindness of Dr. Purves Stewart, we were able to examine sections prepared with a reliable specimen of the stain. Dr. Purves Stewart was also kind enough to look through

FIG. 5.

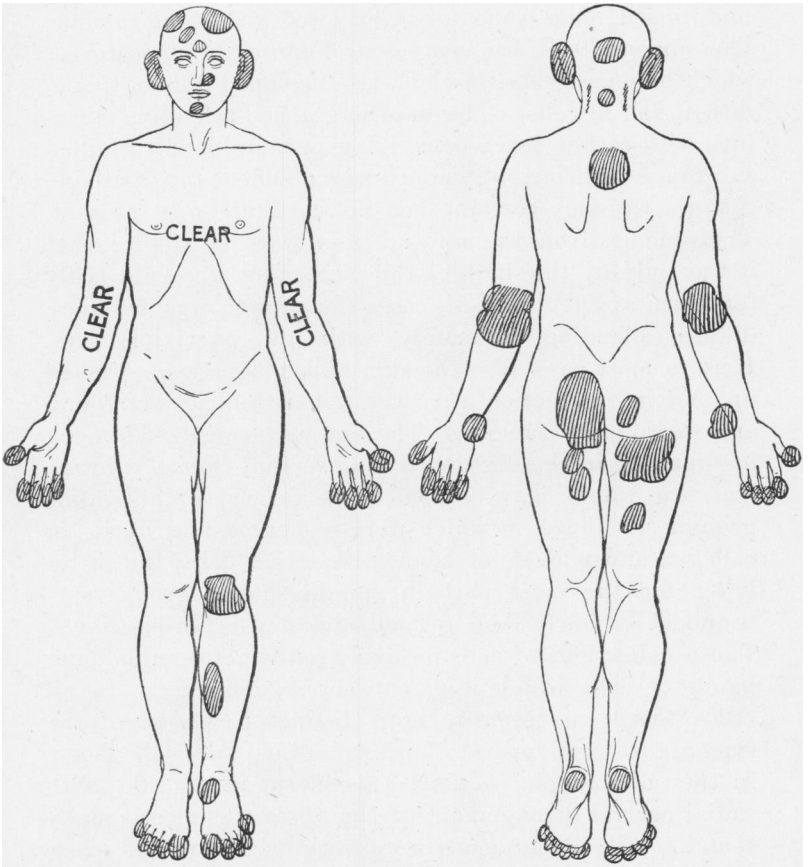


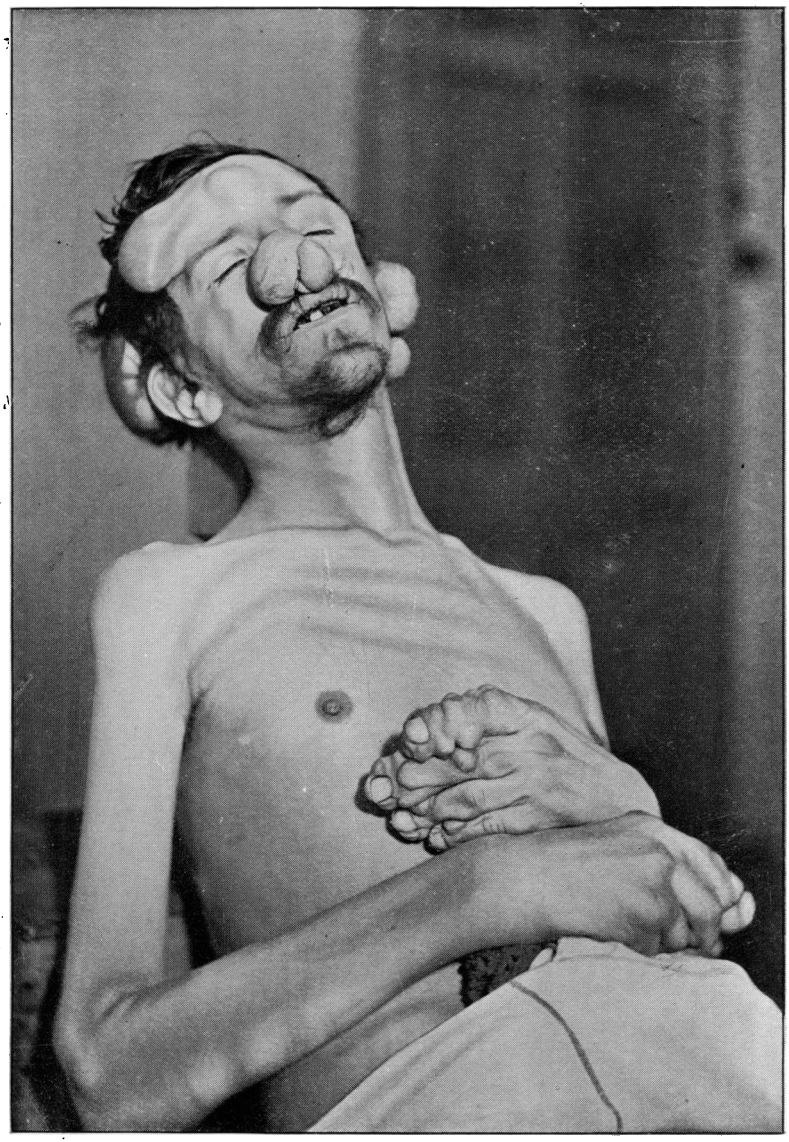
Chart showing distribution of tumours on body and extremities in Ellen.

the sections with one of us, but in no instance was a nerve-fibre found.

Other specimens were stained with a variety of the ordinary histological stains, including that for elastic

tissue, but none was found in any of the tumours. In the case of the finger tumour the growth was found well circumscribed, and only lightly attached to the overlying skin and deeper tissues. All the ordinary appendages of the skin were almost normal, but pushed up and slightly compressed, none being found enclosed within the tumour. The tumour itself was composed of dense fibrous material, which in some parts showed well-developed fibrous tissue, rather rich in cells, while in others it had a hyaline structure with rather large cells, lying in more deeply stained substance bearing a strong resemblance to cartilage, though we feel certain that no cartilage was present. The tumours from the ear and nose were so similar that it was only by the shape of the pieces that we could easily tell them apart. In both cases the growth was far more diffuse, blending intimately with the overlying skin. Here in most cases also the skin appendages were pushed up, but in one section a part of a hair-follicle was found enclosed in the tumour. The fibrous element was much less marked in these growths than in that from the finger, and the whole may be said to be formed of a hyaline ground substance, in which were numerous fine fibrils of collagen and packets of connective-tissue cells, the latter lying for the most part in spindle-shaped groups surrounded by fairly well formed and dense fibrous tissue. These collections of cells probably really represented long bands of cells which were cut across obliquely. In all cases there was seen to be a distinct tendency of the tumours to form separate lobes, and this was well shown in the microscopic sections; a concentric outside with radial central arrangement of the fibres was most usually found. All the tumours were very vascular, but those from the nose and ear especially so, the vessels in every instance being provided with a thin but well-organised wall. The hyaline ground substance was investigated by means of several stains, but without very definitely ascertaining its nature. It was apparently not myxomatous in nature, as it gave no metachromatism with

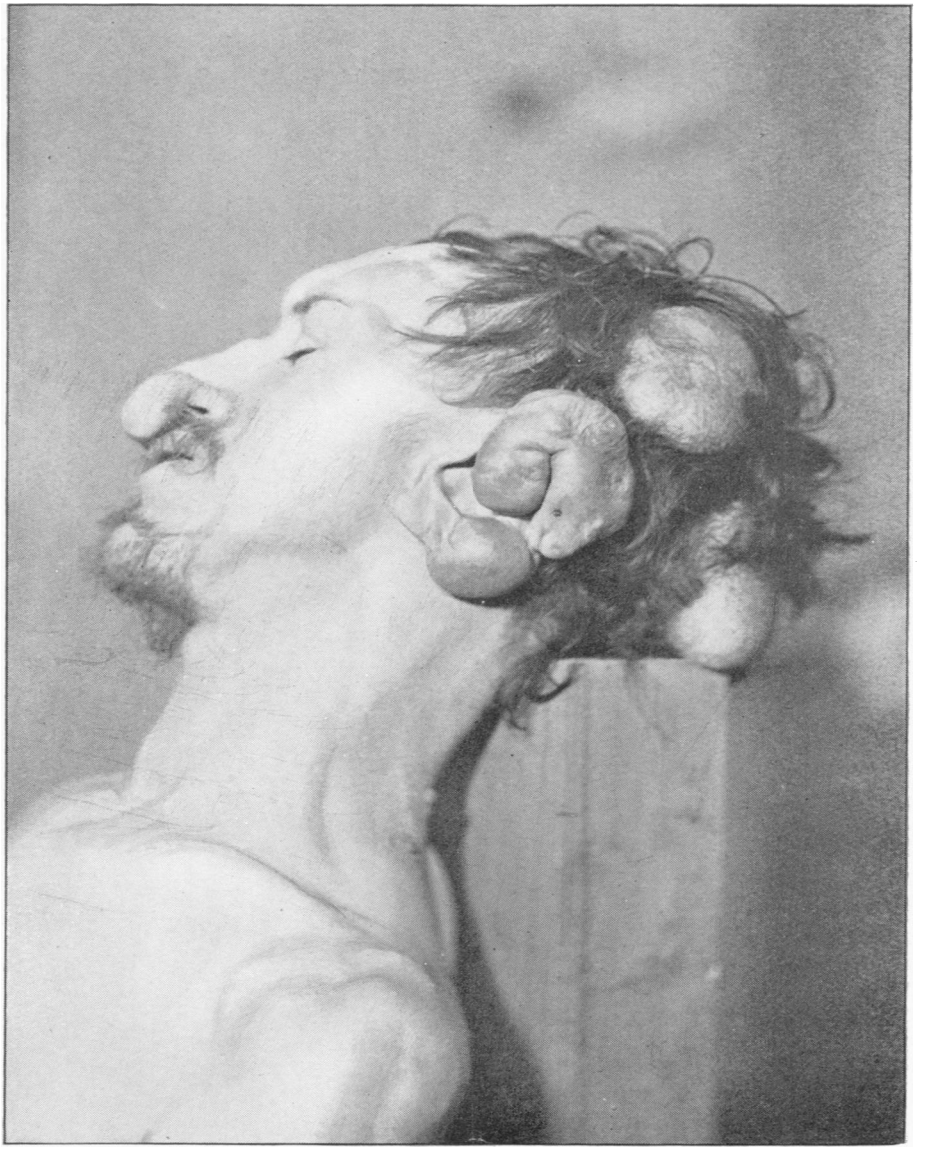
Robinson and Whitfield: Molluscum Fibrosum in Children, Plate I.



PHOTOGRAPH OF RICHARD.
Showing deformity of nose and curious condition of fingers.

Adlard and Son, Imp.

Robinson and Whitfield: Molluscum Fibrosum in Children, Plate II.



PHOTOGRAPH OF HEAD OF RICHARD.
Showing deformity of left ear.

Adlard and Son, Imp.

Subcutaneous node from dorsal surface of the finger.

Bale & Danielsson, Ltd.

Whitfield & Robinson: Molluscum Fibrosum. Plate IV.

Subcutaneous growth from the ear.

Bale & Danielsson, Ltd.

thionin. On the whole the best specimens were obtained with Hansen's modification of Van Gieson's stain. We are quite aware that in making this further report we have not advanced in knowledge of the pathology or etiology of this remarkable disease. We think, however, that it should be taken out of the class of molluscum fibrosum, which is now practically synonymous with neuro-fibromatosis, a disease with which this has apparently nothing in common, and placed simply in the category of multiple fibromata.

DISCUSSION

Sir THOMAS BARLOW had had these cases under his observation for many years after the death of Dr. Murray. In regard to the etiology, Dr. Murray had thought the tumours due to insanitary conditions, but years after the birth of the children the father came under observation with undoubted malignant disease, and died from cancer of the stomach. The eldest of the children was certainly the most severely affected, and the youngest was least, and in her the disease was not progressive, as it was in the former two cases. Each of these children was liable to intercurrent febrile attack, in some cases with increase in the size of the tumours and attacks of dermatitis. In one case, after removal of a tumour surgically, there was very alarming surgical fever. The mental condition of the eldest patient was defective, and there was deafness. The defect was less in the second, and least in the youngest.

Dr. ROBINSON, in reply, said that the mental condition of the patient whom he had seen was similar to that of the eldest sister as described by Sir Thomas Barlow.